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PARAMYOCLONUS MULTIPLEX.

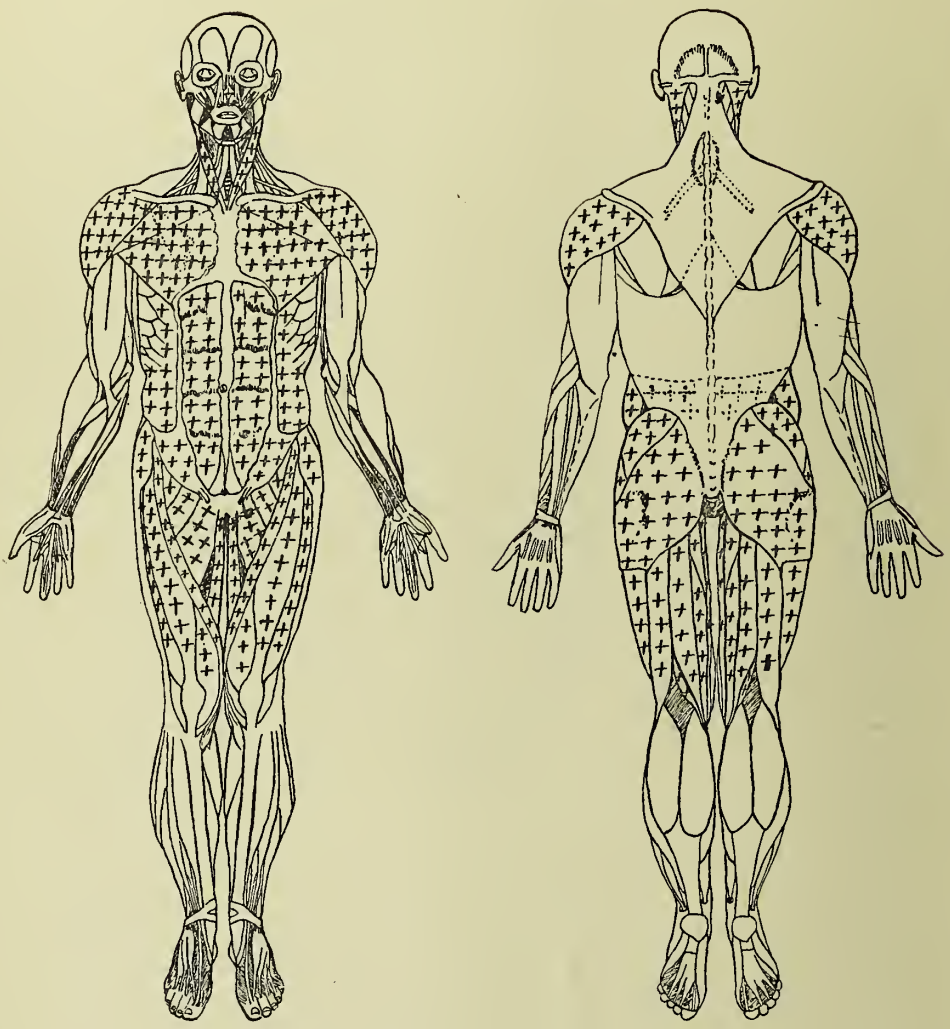
A CLINICAL LECTURE.

BY

F. W. LANGDON, M.D.,

NEUROLOGIST TO THE CINCINNATI HOSPITAL, AND TO THE OPHTHALMIC HOSPITAL OF CINCINNATI;
CLINICAL PROFESSOR OF NERVOUS DISEASES AT MIAMI MEDICAL COLLEGE.

Delivered at the Cincinnati Hospital,
January 28, 1896.



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Distribution of myo-clonic spasm indicated by plus marks.

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Ladies and Gentlemen:

The patient before us to-day presents an excellent example of that rare and interesting malady—*paramyoclonus multiplex;* a name which, though somewhat lengthy, has the merit of conveying an accurate description of the nature and distribution of the leading symptom of the disorder.

We are indebted for this name to Friedreich, who first described the disease in 1886, since which time it has been recognized by clinicians in various parts of the world.

Starr,[1] in 1891, reported a case observed by himself, to which is appended a bibliography up to that date. This is the most complete account at present generally accessible to American readers.

Gowers[2] states that of fifty-two cases recorded up to July, 1895, but thirteen are considered by him as true cases of the disease.

Personally, I have seen but one other case, which was in the service of Dr. Gowers at the "Queen Square" Hospital, London, and furnished the material for the lecture just noted.

Clinically, the disorder is characterized by three groups of symptoms, all of which are fully presented by our patient.

First: Involuntary muscular contractions, shock-like, clonic, bilateral, symmetrical and synchronous.

Secondly: The muscles affected are the intrinsic trunk muscles and those connecting the trunk and extremities.

Thirdly: The contractions occur in *paroxysms* of variable duration (one-half minute to ten minutes), the inter-paroxysmal period varying from a few minutes to a few days.

Negatively, we may note: (1) The face, hands and feet, and usually the forearms and lower leg muscles, are not affected; (2) mentation, motor power and sensory perception are not notably impaired; (3) trophic and vaso-motor symptoms are absent.

The causes of the disease are said to be injury, physical strain, fright, shock and mental worry. Our case appears to be due to the latter cause.

We are indebted to Dr. Mark Brown, our efficient House Physician, for the very careful history of the case which he has just read, the salient points of which are:

The patient, P. H., is forty-eight years old, Irish by birth, married; has six healthy children, ranging from nine to twenty years of age. Occupation, a layer of sewer pipe. He is of good physique, somewhat spare in build, his weight having fallen from 190 to 140 pounds in the past two years. Temperature 98.4°, pulse 80.

He presents an anxious expression of countenance and slight impairment of memory at times; mental condition otherwise good.

Smell and taste are not impaired on either side.

Vision: R. E. $^{20}/_{20}$, L. E. $^{20}/_{200}$.

1 "Familiar Forms of Nervous Disease," N. Y., 1891, p. 244.

2 Clinical Lecture, National Hospital for the Paralyzed and Epileptic, Queen Square, London, July 17, 1895 (not yet published).

* Delivered at the Cincinnati Hospital, January 28, 1896.

Can name and match colors well. Ophthalmoscopic examination, kindly made by my colleague, Dr. C. R. Holmes, reveals blurred disks, evidently due to a former papillitis in both eyes and a posterior staphyloma, probably congenital, in the right eye. The dioptric media are clear.

Visual fields concentrically contracted in both eyes; in the left more than in the right (see chart). No *arcus senilis*.

The pupils are medium in size; respond normally to light, accommodation and convergence, but on convergence he is unable to fix them for more than a few seconds, when some dilatation occurs in both.

Ocular movements are normal.

Nystagmus is absent.

Hearing: Right $^{10}/_{10}$, left $^{4}/_{10}$ (to watch tick). He can recognize the sounds of the Galton whistle up to the highest note in both ears.

Fibrillary tremor of intrinsic tongue muscles.

Other cranial nerves normal.

Motor power of muscles of face. trunk and extremities good and equal, Grasp above the average in power. Right 240, left 240 (lower scale of dynamometer). Three tests were made, and the differences were trifling. The affected muscles, as well as others, respond normally to faradism. Gait normal, except when paroxysm comes on, when it appears "spastic" in character, somewhat like a case of degenerative disease of the pyramidal tracts. Coordination of movement is good in all respects between the paroxysms.

Sensation is not impaired to tact, pain or temperature. The muscular sense and posture sense are good.

Superficial reflexes: Scapular and palmar absent. Epigastric slight. Cremasteric and plantar normal.

Deep reflexes: Jaw-jerk, triceps-jerk and wrist-jerk absent. Marked increase in myotatic irritability of the *pectorales majora* and *deltoides*, equal on the two sides and giving rise in the case of the former muscle to a decided "pectoralis major jerk" of the entire upper extremity when abducted at a right angle to the body and extended over the edge of the cot. Knee-jerks hyper-active and equal. Ankle clonus is absent.

Vaso-motor and trophic disturbances are absent.

The organic reflexes are not impaired.

Pulse 80, small, of high tension; artery felt between beats and somewhat tortuous. Heart examination negative.

Respiration rate 28 between paroxysms; 16 when patient is asleep. During a paroxysm it is costal, irregular and suppressed. A slight friction murmur at left apex. Lungs otherwise normal.

Digestive system: Patient thinks he has "dyspepsia," but a varied diet is apparently digested with comfort, and no abdominal tenderness or tympanites can be detected. Liver and splenic dulness are normal.

Urinary system: Micturition is normal, urine amber-colored, sp. gr. 1027, alkaline. Sediment of phosphates. Albumen and sugar are absent.

Generative system: Sexual desire lessened since illness, but not abolished. Sexual power good; erections normal but frequency lessened.

His family history throws no light upon his illness. He is the first of six children, two of whom died in infancy; the other three are living and well. His father's death, at sixty, was due to accident. His mother died in childbirth. He has no knowledge of "fits" or nervous disease of any kind in his family. He has been an industrious man, well fed, clothed and housed all his life. Is a moderate user of alcohol and tobacco. His previous diseases have been: Measles, chicken-pox and whooping-cough in childhood; typhoid fever at twenty, followed by chronic diarrhea for some months; mumps at thirty-three; attacks of influenza four, three and two years ago, which did not cause him to cease work.

His statement of the events closely preceding his present illness is of much significance. Two years ago, just following his last attack of influenza, he had a guarantee of steady work by the year at fixed and satisfactory wages. A friend persuaded him to resign his desirable situation, form a partnership with him and establish a contracting

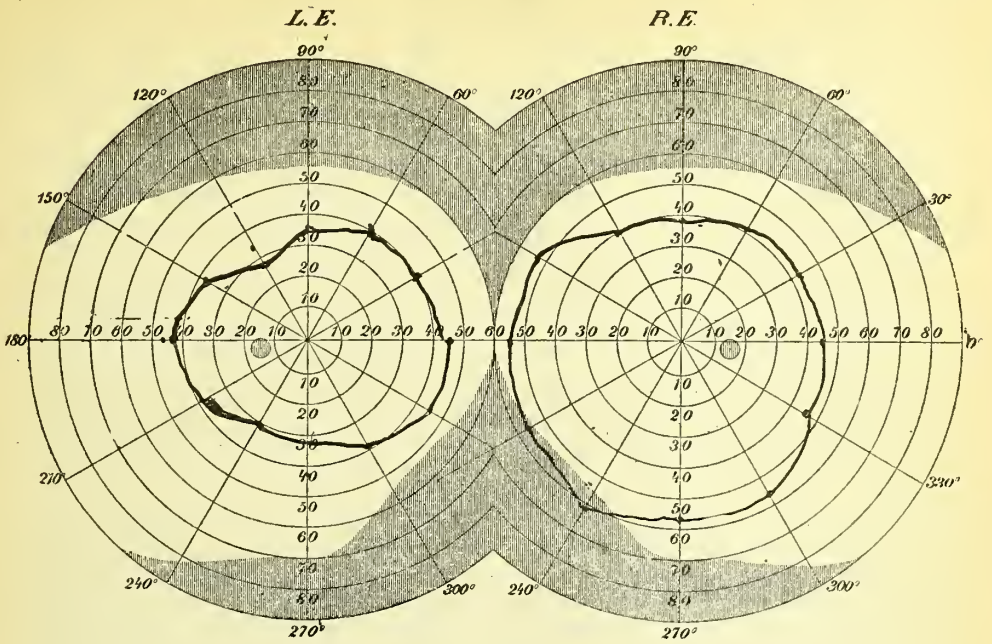

FIG. 1.—Fields for white—good daylight.

business of the same kind on their own account. Ambition, that excellent servant but merciless master, seized upon his soul. He listened to the voice of the tempter and fell. We may surmise, to his credit, that his ambition was of an altruistic form; perhaps he had visions of the six bairns at home, some of whom might yet tread the marble halls of Congress, or even cross the portals of the White House itself. So far as his ambition was concerned, it was as worthy as that of Napoleon himself—even more so.

But these pleasant anticipations were doomed to a sad ending. He tells us that he was greatly "worried in his mind" over the important step he had taken. Two days after resigning his desirable situation, and before entering upon the proposed partnership, he was taken ill with his present malady. No injury, fright, shock, or physical strain of any kind is traceable; and, while the recently preceding attack of influenza was probably a predisposing factor in some degree, yet it is to be noted that it was a mild one, which did not deter him from his work during its progress.

The disease is said to be "often hereditary," which does not appear to be the case here. Males are usually affected, very few cases in women being recorded. Youth and adult life are the usual periods of occurrence. The neurasthenic state probably predisposes.

The onset of his present disease was sudden. While lying in bed he was seized with vomiting, followed in a few moments by the shock-like muscular contractions (which you now notice), and which appeared first in the anterior abdominal wall and in the flexors of the thighs. At first the contractions were even more severe than they now are, and produced violent and repeated flexions at the hips, raising his legs completely from the bed. The duration of these early attacks was about half a minute, and they recurred two or three times a day for two weeks. There was no unconsciousness, no mental confusion, no paralysis. The shock-like clonic contractions continued to recur in paroxysms twice, thrice, many times daily. Their violence mitigated somewhat with time, but they have disabled him so that he has been able to do but about

eight months' work in the two years since they began. The periods during which he was able to do some kinds of work varied from one day to three months.

On his admission to the hospital, three days ago, he presented the conditions already noted. There has been no change of consequence up to the present time. Efforts at concentration of thought on his part seem usually to excite a paroxysm; so also does sudden exposure of the body to the air, sometimes, not always. Voluntary physical effort also tends to the same effect. But voluntary effort to turn and assume the prone position usually arrests a paroxysm if present, as does a deep inspiration at command.

An individual paroxysm observed by Dr. Brown and myself yesterday presented the following features:

Decubitus dorsal. The attack began while the patient was being interrogated respecting some points in the above history. Patient describes an "aura" consisting of a "cold feeling" and sensation like "electricity" in legs, abdomen, chest and arms. This he says often precedes an attack. Onset sudden, with violent, shock-like, clonic contractions of muscles of anterior abdominal wall, the individual segments of the recti standing out prominently during the contraction. The contractions are bilateral, symmetrical and synchronous with similar clonic contractions in the pectorals, cremasters, and the flexors, adductors and extensors of the thighs. The rate of these contractions is seventy for a full minute, but is not rhythmical, the rate varying at different portions of a minute from 50 to 250.

The muscles of the face, neck, upper arms, forearms and hands are not involved; nor are the lower leg and foot muscles, as a rule, though Dr. Brown thinks he has seen the toes flexed and extended on one occasion. During this paroxysm respiration and phonation are irregular and "jerky." The diaphragm is tonically contracted, and respiration is costal. The tongue was protruded at will and kept out indefinitely without spasm during the paroxysm. This paroxysm ceased in four minutes, synchronously with a deep (involuntary) inspiration.

After an interval of five minutes another occurred; then a similar interval and a second recurrence, each of about five minutes' duration.

The paroxysms exhibited now (at intervals of five or ten minutes), as the patient lies before you, correspond with those just described.

In another paroxysm, observed when the patient was in the prone position, the flat abdominal muscles were seen to contract forcibly at their attachment to the lumbar fascia. The quadratus lumborum could be felt to contract at its iliac attachment. These contractions were similar to those noted of other muscles.

At no time have the contractions been observed in the erector spinæ, latissimus dorsi, glutei or trapezius muscles. Occasionally the sterno-mastoids participate in the attack. When a paroxysm occurs while the patient is standing, there is a coarse tremor-like movement of the hands and feet, which, however, is apparently transmitted from the trunk movements, with which it is synchronous. Rarely, at the beginning of a paroxysm, one rectus abdominis contracts alone, but its fellow soon follows. In some slighter paroxysms the cremasters and thigh muscles seem to escape entirely.

The patient has attempted to keep a record of the number of attacks in twenty-four hours, and has noted eleven, but says there were probably four times as many minor attacks not noted. The paroxysms have not been observed during sleep, and he thinks none occur at that time.

He has had occasional vomiting spells with the paroxysms, which seem due to the over-action of the abdominal wall muscles.

The pathology of the disease is unknown. The two autopsies[1] on record revealed no assignable lesion.

The disease is classed, therefore, with the functional neuroses.

Friedreich believed it due to vaso-

1 Schultze, Neurol. Centralblatt, 1886, 363; and Sinkler, "Text-book of Nervous Diseases by American Authors," 1895, 269.

motor spasm caused by mental or physical strain, and producing a hyper-excitability of the brain or cord. Others, among them Starr, are inclined to regard the primary lesion as a peripheral hyper-activity, producing secondary central over-action.

While it may be objected that these views are merely hypothetical, yet it must be conceded that even hypotheses may assist our conception of the relations of facts.

Looking at the present case in this aspect, and accepting the modern doctrine of the neuron of Waldeyer as the anatomical unit in neuro-dynamogenesis, it would appear that abnormal action in any one of six groups of neurons might produce the clinical picture presented. Thus:

1. The primary sensitive neurons, with their bodies in the posterior root-ganglia and their end-tufts at the periphery and in the cord and bulb respectively, may transmit exaggerated afferent impulses to the anterior-horn motor neurons, producing the hyper-kinesis observed.

2. The secondary sensitive neurons of the bulb or cortex may be similarly affected, and the result a hyper-kinesis of cortical origin. We see no evidence, however, that such hyper-excitability exists on the sensitive side. The absence of anæsthesia, hyperæsthesia or paræsthesia to tact, pain and temperature, and of the myo-neuric pains of tabes; of ataxia or other incoördination, is at least presumptive evidence of the integrity of the neurons of the cutaneous and musculo-sensory tracts.

Hyper-activity of the motor neurons, (3) cortical and (4) spinal, may be excluded by (*a*) its rarity without loss of consciousness or organic cause; (*b*) by the absence of motor weakness, tremor and incoördination between the paroxysms; (*c*) by the bilateral character and symmetry of the contractions; (*d*) by the escape of certain of the most highly developed groups, as those of the face, hands and feet. The main sensory and motor paths being thus excluded as the primary seat of pathological action, we must seek elsewhere for the explanation of the symptoms. The existence of special accelerator and inhibitory neurons in the cortex cerebri must be conceded as probable; such mechanisms are comparable to those that influence the frequency of the heart's action. By the development and activity of these mechanisms we may account for the differences in individuals as regards volitional efforts on the one hand and self-control on the other. In other words, one plies the whip, the other puts on the brake. In our case the brake appears to be out of repair, and acts irregularly.

5. An over-action of accelerator mechanisms would account for symptoms here presented; but such over-action is not likely in a man recovering from influenza and mentally depressed from other causes.

6. Defect in function of inhibitory mechanisms dominating the psycho-motor neurons is a condition which physiologically exists in children, hence the frequency of convulsive affections in early life. It is quite conceivable, therefore, that these inhibitory neurons, the latest structures of the cortex in time of development and function, are also, in obedience to ordinary physiological and pathological law, the first to suffer in states of depression. Note, for instance, the petulance of age, and the motor and mental irritability of the neurasthenic.

Here, then, lies the most probable seat of the lesion in our patient.

The peculiar distribution of the muscular spasm, well shown in the accompanying chart,[1] sufficiently excludes a spinal or peripheral origin.

The fact that "associated movements," not anatomical muscle-groups, are involved, points strongly to a cortical seat of the disease.

It is also of great significance that the movements involved in this case are those habitually and most vigorously exercised by the patient at his work of digging and bending alternately for-

1 This very useful chart for recording muscular paralysis, spasm, degeneration, etc., has been recently devised by Dr. W. H. B. Stoddart, of London. To secure uniformity of records I would suggest that plus marks be used for over-action, minus marks for weakness or degeneration, and ciphers for absolute paralysis.

ward and backward in laying pipe. Even the affection of the sterno-mastoids is suggestive here.

To sum up: Both the causal and the symptomatic indications point (1) to the cortex cerebri as the seat of morbid action; (2) that the disorder is a manifestation of defect in neurons or mechanisms having an inhibitory function.

As regards diagnosis, the disease is unmistakable if you have once seen a typical case such as this. The only diseases which resemble it, even superficially, are:

1. Electrical chorea, or Dubini's disease, in which the contractions are shock-like; but here the resemblance ends, for this disease is restricted to a certain district in Italy, begins unilaterally in one arm or leg, is attended with nerve degeneration and muscular atrophy, and is commonly fatal in a few months.

2. Chorea of Sydenham is characterized by the youth of the patient, the non-paroxysmal character of the contractions, their lack of symmetry and synchronism, and the marked involvement of the face, hands and feet.

3. Senile chorea presents less violent contractions, lack of symmetry, and the distribution to the hands and feet characteristic of the choreas.

4. Hysteria differs in its peculiar emotional state, sensory and motor stigmata, and susceptibility to suggestion, all of which are absent in our patient.

The contraction of visual fields which exists is sufficiently accounted for by the organic retinal disease.

The prognosis of the disease is generally favorable; nearly all cases -recover. Death, if it occur, is due to intercurrent or co-existing disease, to which the patient may possess a greater liability by reason of the depressed state which exists.

The indications for treatment are:

1. To sustain the patient, by good diet, fresh air, agreeable surroundings, rest.

2. To mitigate or arrest the paroxysms, for which purpose chloral and other depressants are to be condemned. Of drugs that have been recommended, arsenic and phosphorus seem the most rational.

On theoretical grounds I should think favorably of quinine, for its peculiar effect on the cortical circulation. Strong galvanic currents to the spine have peen recommended and used; it is difficult to see on what grounds if the disease is, as it appears, a cortical one. If electricity is to be used, I should favor the "sinusoidal" current of high potential, rapid alternation and small quantity, to the head and spine for the marked nutritional effects which seem to follow its use in other functional and nutritional neuroses.

I shall order for this patient quinine in moderate doses, increased till its physiological effects are obtained. Should that fail to influence favorably the disease, I shall resort to the sinusoidal current. In either event I hope to report to you the results of treatment later in the course.

Addendum: Under the quinine treatment for three days, the paroxysms have lessened in number from twenty or more per day to two, and those are comparatively mild in character.